Congenital inguinal hernia, hydrocoele and undescended testis

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Abstract

Congenital inguinal hernias (CIH), hydrocoeles and undescended testes (UDT) are common groin conditions in neonates, infants and children that are encountered by general practitioners, paediatricians, general surgeons and paediatric surgeons. CIH, hydrocoeles and UDT share a common embryological origin. Clinical differentiation between the three conditions can be challenging, particularly as they may exist in isolation or combination in the same patient. Accurate clinical distinction is imperative as the management and outcome is different for each condition. Surgery and outcomes for these conditions is discussed.

Keywords Children; congenital inguinal hernia; hydrocoele; undescended testis

Congenital inguinal hernia

Definition

A congenital inguinal hernia (CIH) is an indirect hernia related to failure of closure of the patent processus vaginalis (PPV) at the deep inguinal ring. Intra-abdominal contents pass within a PPV, through the deep inguinal ring, inguinal canal, superficial inguinal ring and potentially into the scrotum (male) or via the canal of Nuck to the labium (female).

Incidence and age at presentation

The incidence of congenital inguinal hernia depends on age. The incidence is highest in premature infants. The incidence may be as high as 60% in infants born at 500–750 g. Nearly one-third of infants with a birth weight less than 1000 g will develop an IH. Term infants have an IH incidence of 3–5%.

The overall incidence of childhood inguinal hernia is 0.8–4.4%. Inguinal hernia repair is considered the most frequently performed paediatric surgical operation.

Sex and laterality

Congenital inguinal hernias are between four and ten times more common in boys than girls. In boys, CIH occur on the right in 60–70%, left 20–25% and bilaterally in 5–15%. Laterality is equal in girls.

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Family history and geography

A higher familial incidence has been reported, although, unlike umbilical hernias, incidence does not seem to vary with ethnicity or geographical region.

Aetiology

Inguinal hernia is known to be associated with certain conditions. These include connective tissue disorders (cutis laxa, Ehlers–Danlos syndrome, prune belly syndrome), ascitic conditions (ventriculo-peritoneal shunts, peritoneal dialysis, chylous ascites, biliary atresia), other conditions causing increased intra-abdominal pressure (post closure of gastroschisis and exomphalos, cystic fibrosis) and in association with other developmental sequelae (undescended testis).

Embryology and pathology

Congenital inguinal hernias, hydrocoeles and undescended testes are linked to the descent of the gonads and subsequent closure of the processus vaginalis. Much of the knowledge regarding the detailed mechanisms of testicular descent and the role of the processus vaginalis has been elucidated by the research team of Professor John Hutson in Melbourne, Australia and reading their findings is recommended for a detailed understanding of the embryology of the testes and groin.

Essentially, the gonads are derived from coeliac epithelium and primordial germ cells, and develop on the urogenital ridge on the posterior abdominal wall. Until the 6th week of gestation, male and female gonads are indistinguishable. During the 7th week of development the testis starts to differentiate. The gubernaculum arises around this time, originating at the testis and inserting into the anterior abdominal wall at the site of the future inguinal canal. As the fetal abdomen enlarges, from the 8th to the 15th week, the testis is held at the level of the internal ring, while the mesonephros (which will become the kidney) ascends to its eventual retroperitoneal position. At around 12 weeks a fold of peritoneum starts to protrude towards the internal ring. This finger-like diverticulum is the processus vaginalis, on which the testis slides through the inguinal canal. The processus vaginalis lengthens through the inguinal canal from the 3rd to 7th month of development, and the gubernaculum shortens, drawing the testis downwards. Between 36 and 40 weeks, the testis reaches the scrotum and the processus vaginalis gradually obliterates, closing the peritoneal opening at the internal ring. Just the very distal portion of the processus vaginalis remains, sitting around the testis as the tunica vaginalis.

Failure of obliteration of the processus vaginalis at the level of the internal ring (a patent processus vaginalis) is observed in congenital inguinal hernia and hydrocoele (Figure 1). Closure occurs earlier on the left than the right and this may be related to the higher incidence of right-sided hydrocoeles, hernias and undescended testis.

At birth, up to 95% of male infants can be observed to have a patent processus vaginalis. Obliteration of the PPV occurs in the postnatal period, so that at 1 year the incidence of PPV is 40% and 20% at 2 years. The relationship between obliteration or closure of the PPV and development of CIH is not yet fully understood.

Presentation and clinical features

The classic clinical presentation of a congenital inguinal hernia is an intermittent bulge in the groin, scrotum or labia. This is most visible when the child is crying, coughing or performing other manoeuvres that increase intra-abdominal pressure (e.g. laughing). Older children may complain of vague ache in the inguinal region. On examination, there may be an appreciable bulge in the groin superiomedially to the pubic tubercle, but often the hernia is reduced at rest. Provocative manoeuvres (such as standing them up, coughing, making the child laugh or jump) are required to elicit it. The 'silk glove sign' has traditionally been used as supporting evidence of an inguinal hernia: on placing a single finger over the cord and gently rolling it over the pubic tubercle there is a palpable thickening and the sensation of rubbing two pieces of silk together. This has a sensitivity of 93% and a specificity of 97%. Increasingly, parents are able to provide clinicians with photographic evidence of the hernia bulge through the use of handheld digital devices. Examination should include the scrotum to confirm testicular position, as an undescended testis may present as a groin mass.

A hernia may become incarcerated (irreducible) which results in bowel obstruction or strangulation (loss of blood supply) of the hernial contents. An incarcerated hernia presents with a hard, tender groin swelling which cannot be reduced with gentle pressure. Symptoms of obstruction such as vomiting and abdominal distension may be present. Severe pain, prolonged

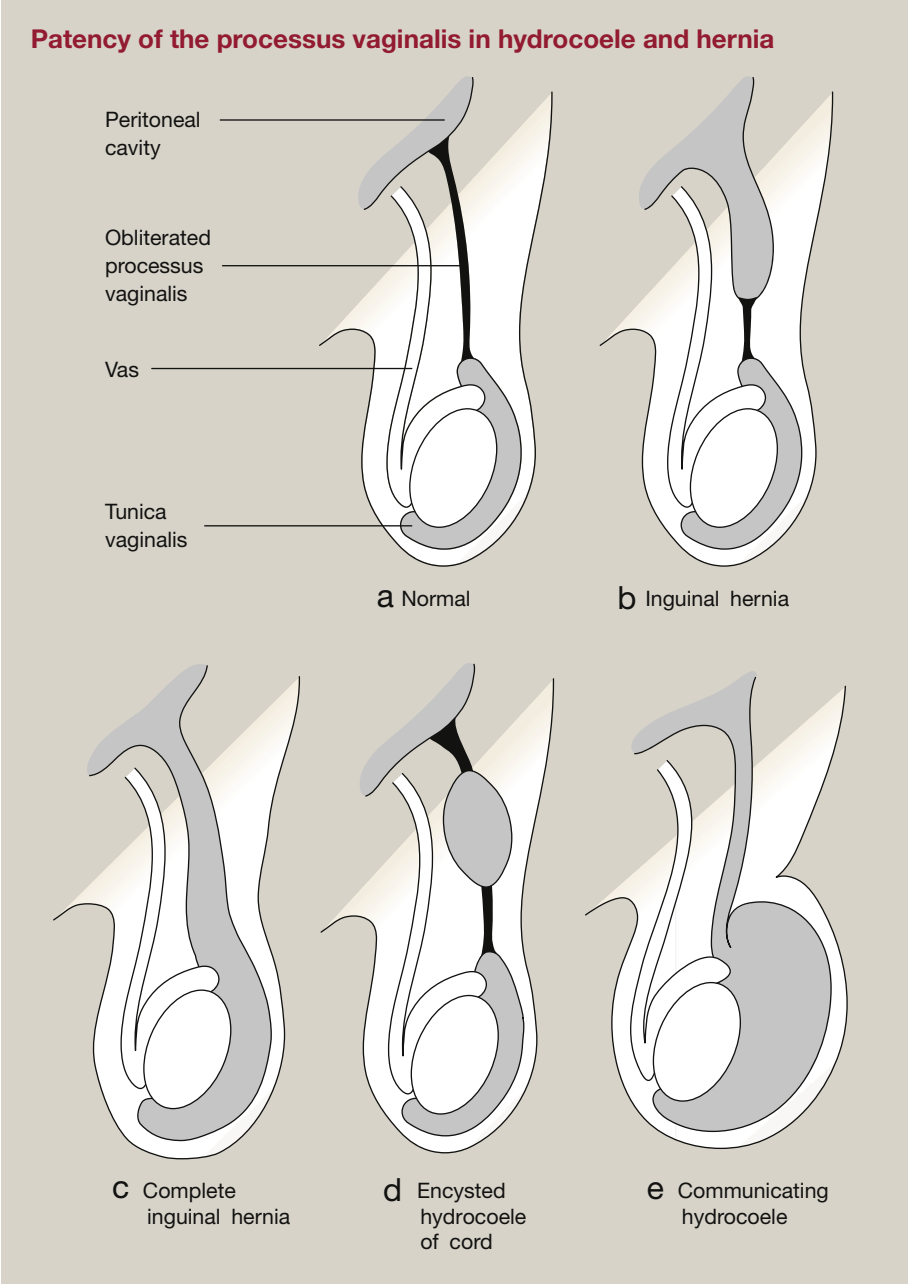

Figure 1

history of incarceration, fever, tachycardia and vomiting are suggestive of strangulation. Unfortunately, up to half of infants with a CIH present with an episode of incarceration. Incarceration necessitates immediate resuscitation and attempted reduction or prompt surgery if reduction fails. In practice, the majority of indirect hernias are reducible if performed by an experienced clinician.

Hernial sac contents

A hernial sac may contain small bowel, ovary, omentum, appendix (Amyand's hernia 1735), a Meckel's diverticulum (Littre's hernia 1700), meconium (seen in neonatal intestinal perforation as green tinged staining of the inguinal canal or scrotum) or tubing (ventriculo-peritoneal shunt or peritoneal dialysis catheter). A hernia sac containing appendix, ovary or omentum may explain failure to reduce a hernia.

Differential diagnosis

Differentiation between an irreducible inguinal hernia, a hydrocoele and torted testis in an infant can be difficult. The infant is often hungry, irritable and crying. If the groin swelling has been present for more than a few hours oedema makes anatomical landmark identification difficult. A hydrocoele can usually be identified by the ability to 'get above' the swelling when palpating with index finger and thumb in the direction of cord structures towards the abdomen (except in the rare case of an abdominoscrotal hydrocoele). Transillumination is not a reliable sign of a hydrocoele in an infant, as a hernia may also transilluminate. A reducible groin lump is indicative of an inguinal hernia as long as the testis remains palpable in the scrotum. It is possible to mistake an emergent testis (an undescended testis that passes between abdominal cavity to inguinal canal) for an inguinal hernia if the testis has not been identified separately. An infant with a torted testis and discoloured oedematous scrotum that extends into the groin can be clinically indistinguishable from an incarcerated inguinal hernia.

If doubt exists, urgent inguinoscrotal ultrasound should be able to visualize bowel contents passing from the abdominal cavity into the inguinal canal confirming an incarcerated hernia that needs either immediate reduction or urgent groin exploration. Ultrasound will not be able to fully differentiate between a hydrocoele and a torted testis with reactive hydrocoele. If the possibility of a torted testis exists, urgent scrotal exploration is necessary. Ultrasound should prevent an unnecessary scrotal exploration finding of small bowel in the scrotum if an incarcerated inguinal hernia has been mistaken for an acute scrotum.

The differential diagnosis of groin swelling in a child is shown in Table 1.

Differential diagnosis of a groin or scrotal swelling

	Differential diagnosis	Cardinal features
Reducible groin mass	Reducible inguinal hernia	Non-tender, above and medial to pubic tubercle, soft, reduces
	Communicating hydrocoele	Non-tender, palpable fluid, transilluminable
	Femoral hernia	
Irreducible groin mass	Incarcerated inguinal hernia	Pain, hard, obstructive symptoms
	Incarcerated femoral hernia	Pain, hard, obstructive symptoms
	Non-communicating hydrocoele	Non-tender, transilluminable location
	Enlarged inguinal lymph node	Defined mass, may be possible to milk into scrotum. If presents with pain, consider torsion of UDT
	Undescended testis	
Scrotal mass	Inguino-scrotal hernia	Mass contiguous with groin 'cannot get above it' may be reducible or incarcerated, as above
	Torsion of the testis	Testicular pain, swelling confined to scrotum 'able to get above'
	Torted hydatid of Morgagni	As above, may see 'blue dot'
	Idiopathic scrotal oedema	Bilateral, painless, often preceded by viral illness
	Testicular/scrotal malignancy	

Table 1

Management

Assuming the diagnosis is correct, an inguinal hernia will not resolve spontaneously and surgery is necessary to treat this condition. As soon as an inguinal hernia is diagnosed in a young infant, referral for surgery should be made and surgery should occur within weeks to minimize the risk of incarceration. While some general surgeons with appropriate training in the general surgery of childhood perform inguinal herniotomies in older children, neonatal herniotomy should always be performed by paediatric surgeons.

Congenital inguinal hernia surgery can be performed open or laparoscopically. The key components of the procedure involve: ensuring the hernia sac is emptied of its contents, preserving the integrity of the cord structures (particularly vas deferens, testicular vessels and ilio-inguinal nerve), closure of the hernia sac, and ensuring that the testis is located at the floor of the scrotum at the end of the operation.

Open inguinal herniotomy – males: It is important to ensure that the hernia is reduced before commencing surgery as this makes the operation safer and less technically challenging.

Incision: A consistent groin skin crease exists in neonates, infants and children and this usually represents the optimal site for incision. Scarpa's fascia is opened and dissection through a variable amount of adipose tissue will reveal external oblique and the inguinal ligament. The key to safe inguinal herniotomy is adequate exposure of the inguinal ligament, superficial (external) ring and the cord as it emerges through the superficial ring. Gentle traction on the testis will aid identification of the cord structures as the cord will be seen to move in the inguinal canal. The fibres of external oblique can be seen to decussate around the superficial inguinal ring. Usually the external oblique/inguinal canal is opened with a small scapel incision and the cord then delivered. Gentle traction on the cord and blunt dissection

will reveal an inverted 'V' tissue plane that permits the passage of a clip to isolate the cord. The operator should remember that the femoral vein lies deep to the inguinal canal and the urinary bladder medial to the inguinal canal. Once the cord is isolated the hernial sac (generally white in colour) is grasped with forceps and the cremaster muscle and fascia is swept off the sac. When in the correct tissue plane this manoeuvre proceeds to reveal first the testicular vessels and then the vas deferens. The vessels and vas must be treated with due respect and never held with a forceps for fear of causing testicular atrophy or occlusion of the vas. The vas is identified by its white colour and fine blood vessels on its surface, but its location is confirmed by rolling it between index finger and thumb, which enables the cord-like thickness to be appreciated. Once the testicular vessels and vas have been isolated, if there is any doubt as to whether sac is empty of not, the sac can be opened between clips to confirm that the sac is empty and the sac can be clipped. The vessels and vas are dissected off the sac towards the deep (internal) ring so that the sac can be twisted and transfixed with a single or double absorbable suture. Care should be taken when separating the vessels and vas from the sac towards the deep ring as the sac can be extremely thin at this point. An unappreciated defect in the sac at this point may lead to recurrence and probably explains the high recurrence rate in neonates. Gentle pressure on the abdomen allows a small amount of peritoneal fluid to bulge into the proximal sac and confirm that it is intact.

The external oblique, Scarpa's fascia and skin are closed in layers using absorbable sutures. The testis is confirmed and documented to be in the scrotum at the end of the procedure.

The patient is allowed to drink and eat when awake and discharged home the same day unless they are a neonate. Routine follow up for straightforward herniotomy is not usually necessary, but is important if the hernia has been strangulated to monitor for testicular atrophy.

Open inguinal herniotomy – females: Herniotomy in girls does not present the same challenge as boys, as neither the vas deferens nor testicular vessels are present. The hernia sac in girls may contain ovary, which may not be reducible. An ovary may undergo torsion in a hernial sac and urgent surgery should be considered to allow preservation of the ovary. The hernial sac is usually dissected out in its entirety transfixed at the deep ring and excised. The deep ring is usually closed at the end of the procedure in girls, which helps to prevent hernia recurrence.

Laparoscopic herniotomy: A port for the camera is placed at the umbilicus and instruments are passed via stab incisions or ports either side of the umbilicus. The open deep inguinal ring is identified. A purse-string suture is passed, catching the peritoneum and avoiding the vas and vessels circumferentially. As the purse-string is tightened the open ring is seen to be closed. One or two sutures may be necessary.

Complications of herniotomy

Complications include wound infection, bleeding, testicular atrophy (1% elective and up to 30% if incarcerated), injury to vas deferens (1% – repair with fine suture 8/0), recurrence (1% in older children; 4–8% in neonates), and iatrogenic ascent of testis.

Pitfalls

1. Metachronous hernia (contralateral hernia that appears after the ipsilateral hernia has been repaired) occurs in approximately 1 in 14 patients and generally this risk is not thought to be high enough to warrant routine contralateral exploration.
2. Risk of androgen insensitivity in girls presenting with inguinal hernia. Approximately 1% of girls that present with an inguinal hernia will have androgen insensitivity and XY karyotype. Approaches to this small but significant risk vary and include routine counselling and assessment of karyotype.

Congenital hydrocoele

A congenital hydrocoele is an abnormal collection of fluid in the processus vaginalis which fails to obliterate resulting in a patent processus vaginalis (PPV) and swelling in the scrotum or groin.

Incidence and sex

Approximately 5% of boys born at term have a hydrocoele and by the age of 2, 90% of these will have resolved. Hydrocoeles are less common in girls and known as a hydrocoele of the canal of Nuck.

Aetiology

Hydrocoeles may be congenital or acquired. The aetiology of congenital hydrocoeles has been outlined above. Acquired causes are: trauma, tumour (lymphoma, teratoma, rhabdomyosarcoma), infection (viral or bacterial) or secondary to torsion of the testis or hydatid of Morgagni, or iatrogenic following surgery for hernia or varicocoele.

Clinical features

A scrotal or groin swelling is usually noticed by the parents and is often seen to increase in size following an intercurrent viral infection. The overlying skin is often noted to have a blue tinge. Sometimes the hydrocoele is tense and it is difficult to palpate the testis separately. Unless the hydrocoele is a rare abdominoscrotal variant, the examiner should be able to 'get above' the swelling. Hydrocoeles transilluminate, but this is only a safe discriminator in older children. Green/blue discolouration of the groin or scrotum of a neonate may indicate an intestinal perforation with meconium passing into a PPV.

Investigation

The diagnosis of a hydrocoele is usually made on clinical examination. If the testis is abnormal to palpation, or the hydrocoele has unusual features, an ultrasound will help exclude a possible tumour.

Management

The arbitrary age at which many paediatric surgeons offer surgery to ligate a patent processus vaginalis is 2 years of age. The rationale being that after the age of two, spontaneous resolution is unlikely.

Surgery: ligation of patent processus vaginalis: Surgery is similar to herniotomy. The PPV is either fluid filled or a thin white structure related to the testicular vessels and vas with the

same orientation as the sac in an inguinal hernia. The PPV is safely isolated and transfixed proximally. Distally the PPV is opened and the hydrocoele fluid expressed, so that the scrotal swelling disappears. Complications are similar to those for inguinal herniotomy and are rare. Adolescents may have a non-communicating hydrocoele and require a scrotal hydrocoelectomy or plication (Jaboulet or Lord procedure).

Undescended testis

Incidence

The incidence of undescended testis (UDT) is 1% in a term newborn male.

Management

The British Association of Paediatric Urologist (BAPU) released a Consensus Statement on Undescended Testis in 2011. A NICE clinical knowledge summary was updated in 2014. There has been a gradual trend in performing orchidopexy on children at a younger age. Current guidance is that referral should occur at 3 months with the aim of orchidopexy being performed between 3 and 12 months of age.

Indications for surgery

The following are all cited as reasons for performing an orchidopexy:

- To place testis in an examinable position to detect malignancy early.
- To potentially reduce malignancy rates.
- To optimize fertility.
- To reduce risk of torsion of UDT.
- To reduce risk of trauma.
- To optimize hormonal function.
- To reduce possible discomfort.
- For cosmetic and psychological reasons.

Studying outcomes in UDT has significant methodological difficulties, including the long time (20–40 years) between intervention (orchidopexy) and outcome (malignancy or fertility) and also difficulties confirming fatherhood. Scandinavian longitudinal studies have provided some invaluable evidence. Overall, the risk of testicular malignancy is estimated to be three to ten times the background risk, depending on relative age at orchidopexy and whether the testis is intra-abdominal. Although contentious, Petterssen has demonstrated a 2.23 times relative risk of testicular malignancy if orchidopexy is performed under the age of 13, compared to a 5.40 times relative risk if orchidopexy is performed over the age of 13 years.

Fertility varies between 87.5% and 14% depending on age at orchidopexy, site of testis and single or bilateral pathology. Laboratory work by Hutson has suggested that orchidopexy should occur at 3 months to optimize normal spermatogonia development, although this has to be weighed against the potential increased risks of damage to cord structures, etc. during orchidopexy in younger infants.

Surgery

Palpable undescended testis: The testis is confirmed to be present in the groin – either preoperatively or during examination under anaesthetic. Surgery is similar to herniotomy and involves groin skin crease incision (usually slightly larger) and an additional scrotal incision. The cord is delivered, the testis located and the gubernaculum divided. The testicular vessels and vas deferens are separated from the processus vaginalis (PV). The PV is usually diaphanous, particularly at the point where the vas and vessels are attached, necessitating gentle and patient separation of PV from the vas and vessels to preserve the integrity of the PV. The PV is divided and transfixed at the deep ring. Dividing the PV allows the testis to move caudally to its ultimate goal, the floor of the scrotum. The mobility of the testis is limited by length of the testicular vessels. Manoeuvres useful in mobilizing a testis under tension to the scrotum include: dividing 'lateral bands' of tissue around vas and vessels, skeletonizing the vas and vessels so that the vas and vessels are clearly seen to move away from each other as their paths are traced into the retroperitoneum. A transverse scrotal incision allows the retrograde passage of a clip that then pulls the testis via the groin into the scrotum. It is important to ensure that the vas and vessels are not twisted to prevent testicular loss. The testis is placed in a subdartos pouch and it may also be anchored in place with an absorbable suture. Wound closure is as per herniotomy. The testis is confirmed to be in the scrotum at the end of the procedure. Orchidopexy is usually performed as a daycase surgery.

Follow-up should ideally be carried out until a year after the operation to confirm the survival and position of the testis.

Impalpable testis: If a testis is impalpable in outpatients, examination under anaesthetic is performed. If the testis remains impalpable, laparoscopy can then be performed to locate the position or absence of the testis. Standard laparoscopy using a 5-mm umbilical optical port and 5-mm working ports is carried out. At laparoscopy the surgeon may find:

- An intra-abdominal testis – the surgeon then needs to determine whether single or two stage orchidopexy, or orchidectomy is required.
- An absent testis – visible atrophic vas and vessels but no visible testicular tissue.
- The vas and vessels entering the deep ring – a groin exploration should then be carried out to look for a missed 'impalpable testis' or an atrophic testis/testicular remnant. The former may contain potentially viable tissue, but a small remnant should be removed as it offers no benefit and potentially contains oncogenic testicular tissue.

If an intra-abdominal testis reaches the contralateral deep ring, single-stage orchidopexy is deemed to be possible. If, despite peritoneal mobilization, this is not possible, then a two-stage orchidopexy is required (Fowler–Stephens procedure). The first stage involves ligation of the testicular vessels. The testis is usually able to survive on the blood supply to the vas alone. The second stage occurs 6 months later and uses laparoscopy to assess survival of the testis before the testis is placed within the scrotum.

Complications of surgery

Complications include wound infection, bleeding (particularly of the testicular vessels), testicular atrophy (5–25% single versus two stage), injury to vas deferens (1–5%) and re-ascent of the testis (5%). ◆

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